calculations can be used to differentiate patients with large, clinically significant left-to-right shunts from those with insignificant ones. We have found these techniques, while time-consuming, to be helpful in the serial management of patients.

The second Doppler method involves the prediction of transvalve gradients. The method is based on the Bernoulli principle in which the potential energy lost as blood goes from a high pressure area to a low pressure one across an obstruction is changed into kinetic energy. The velocity of erythrocytes is substantially higher downstream from the obstruction than it is proximal to the stenotic area. In a case of aortic stenosis, high velocity flow is detected and sampled in the ascending aorta. In pulmonary stenosis, high velocity occurs in the main pulmonary artery and is sampled under two-dimensional echo guidance. We and other investigators have found that this technique is simple to apply and yields gradient estimations with an accuracy of within 5% of measurements obtained at cardiac catheterization in cases of both aortic and pulmonary valve stenosis. Certainly, in pulmonary stenosis the method obviates the need for cardiac catheterization before submitting patients to a surgical procedure. In a case of simple aortic stenosis with normal ventricular function, cardiac catheterization can also be avoided. The gradient method has proved to be of substantial importance for serial study of patients as they grow because aortic stenosis of childhood is often a progressive disease.

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Monitoring for the Sudden Infant Death Syndrome

THE SUDDEN INFANT DEATH SYNDROME is a sudden and unexplained death, usually occurring between 1 month and 1 year of life. Sudden infant death syndrome accounts for about 2 deaths per 1,000 live births in the United States and represents the leading cause of infant mortality after the neonatal period. If there is enough evidence to explain the death, "sudden infant death due to _____" should be designated, which is distinct from the sudden infant death syndrome. The risk for sudden infant death is increased in neonatal survivors who were born prematurely and in infants born to opiate-addicted mothers. Compared with the overall incidence of about 0.2%, the risk for sudden infant death syndrome has been reported to be from 0.56% to 2% in subsequent siblings and 4% in a surviving twin.

Infants presumed to be "healthy" who have a "life-threatening event" may have received vigorous stimulation, mouth-to-mouth breathing or cardiopulmonary resuscitation from the parents or a babysitter. Some have referred to these infants as "near miss" for the sudden infant death syndrome when there is no specific diagnosis, but the more preferred term may be "apnea of infancy."

Knowledge regarding the cause and optimal management of prolonged apnea (cessation of breathing for 20 seconds or longer or a shorter respiratory pause associated with bradycardia, cyanosis or pallor) and its relationship to the sudden infant death syndrome is still limited. Most infants who have apnea of infancy survive, although the risk of sudden infant death occurring in this group is somewhat greater than in the general population. Many infants who have prolonged apnea are perceived by parents and physicians as having had a life-threatening event that may recur. The medical evaluation of these infants includes a careful history, physical examination, electronic cardiorespiratory monitoring and other appropriate studies done in hospital to determine the cause and severity. Apnea may be associated with such conditions as infections, metabolic aberrations, seizure, cardiac arrhythmias or congenital heart disease, airway obstruction, gastroesophageal reflux and impaired regulation of breathing.

In most cases, the pediatric evaluation and cardiorespiratory electronic monitoring can be initiated by a community pediatrician if these resources are available in a community hospital. In general, initial studies that may be helpful include a complete blood and differential count, arterial blood gas determination and chest radiograph. An electrocardiogram may be helpful if there are clinical findings suggesting arrhythmia, heart disease or pulmonary hypertension. If the history suggests seizure as a possibility, an electroencephalogram may be helpful. Other studies may be suggested by the clinical presentation and examinations. Appropriate treatment for a specific cause will often resolve the apnea problem.

When there is no diagnosis and the infant is asymptomatic, the risk for a recurrent episode of life-threatening prolonged apnea is unknown and home impedance apnea-cardiac monitoring may be used to detect prolonged apnea and alert the family to a possible "life-threatening episode." There is currently no single test or group of tests available to determine risk of prolonged apnea or the sudden infant death syndrome in an individual infant. Clinical follow-up should be arranged with the private medical doctor. Parents should keep records of monitor alarms, observations of the infant during an alarm and the type of intervention used. Consultation with a tertiary center is recommended if an infant continues to have unexplained prolonged apnea in hospital or if an infant, after discharge home, has recurrent apnea that requires vigorous stimulation or

resuscitation. About 40% of infants with apnea of infancy will not have a recurrence.

Management of asymptomatic infants who may be considered epidemiologically at increased risk for the sudden infant death syndrome is controversial. Home monitoring of other asymptomatic infants is not recommended as there are no reliable tests that predict an infant at risk for prolonged apnea or sudden infant death.

Home apnea-cardiac monitors and the alarm limits must be prescribed by an attending physician and should be continued until judged no longer appropriate. Most centers recommend impedance-type apnea-cardiac monitors with alarm limits for apnea and bradycardia. Parents and others involved in an infant's care must be taught monitor use and also cardiopulmonary resuscitation. Social support that includes trained relief personnel and 24-hour equipment service should be available.

Home monitoring may be discontinued when (1) an infant has had no "life-threatening" events for at least three months, or for two months if there have been no problems since the presenting episode; (2) the infant has not experienced a real monitor alarm for at least two months (at an apnea setting of 20 seconds and heart rate setting of 60 beats per minute); (3) during the asymptomatic period the infant must have had an upper respiratory tract infection, an immunization or another illness without recurrence of symptoms; (4) clinical evaluations that include neurologic, developmental and physical examinations show that the problems or initial reasons for monitoring have resolved, and (5) the infant has shown no abnormalities on cardiorespiratory recordings if those were present at the time of the infant's initial evaluation.

The etiology and optimal management of apnea of infancy is not clear and the actual relationship between apnea and the sudden infant death syndrome has not been established. Continued research in these areas is essential.

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Management of Congenital Hypothyroidism

IN 1980 California began a newborn screening program for cases of congenital hypothyroidism. On filter-paper blood specimens, the serum thyroxine (T_4) level is measured followed by confirmatory thyroid-stimulating hormone (thyrotropin) levels on those samples with low T_4 values. The incidence of congenital hypothyroidism is about 1 per 3,750 live births and is confirmed by low T_4 and raised thyrotropin levels.

In October 1983 the California Newborn Screening Program began reporting as abnormal only those blood values that are presumptively positive for primary hypothyroidism—that is, blood specimens that showed low T_4 and raised thyrotropin values, indicating malfunction of the thyroid gland. Although relatively uncommon, other abnormalities such as low T_4 and low thyrotropin levels will be reported as normal for primary hypothyroidism. The most common disorders in which low T_4 and low thyrotropin levels occur include transient hypothyroxinemia, thyroxine-binding globulin deficiency and cases of premature infants and other sick newborns who have sick euthyroid syndrome. Thyroid disorders other than primary hypothyroidism, which include pituitary thyrotropin deficiency (secondary hypothyroidism) or hypothalamic thyrotropin-releasing hormone deficiency (tertiary hypothyroidism) and compensated hypothyroidism (normal T_4 and raised thyrotropin levels), are also now reported as normal for primary hypothyroidism. A diagnosis of primary congenital hypothyroidism should be confirmed by repeating serum T_4 and thyrotropin determinations in a local laboratory. The primary physician must not rely on the state's screening program when there may be a thyroid disorder other than primary hypothyroidism. Thyroid function tests should be done in a local laboratory whenever clinically indicated.

Once a primary physician has been notified that primary hypothyroidism may be present, confirmatory T_4, triiodothyronine resin uptake and thyrotropin levels should be determined in a local laboratory. A thyroid scan using either sodium pertechnetate Tc 99m or sodium iodide I 123 is helpful in determining abnormal development of the thyroid gland. An x-ray film of the knee and ankle for skeletal maturation often shows delayed development and is helpful in determining the duration of hypothyroidism.

Therapy for congenital hypothyroidism should consist of levothyroxine sodium (Synthroid), 8 to 10 μg per kg of body weight a day. With advancing age the dose of levothyroxine decreases to 4 to 6 μg per kg a day in 1- to 5-year-old children and 3 to 4 μg per kg a day in adolescents. Dosages of levothyroxine must be individualized to keep serum T_4 levels in the normal range for age. T_4 levels normally peak at 1 to 3 days of age and reach values of 12 to 22 μg per dl. T_4 levels in preterm infants are lower than those in term infants. After 2 weeks of age and throughout infancy and early childhood, the T_4 levels are about 7 to 15 μg per dl. The T_4 level then gradually decreases to 4.5 to 12.5 μg per dl after 15 years of age, when the normal adult levels are reached.

With inadequate thyroid hormone replacement, growth and mental development may be impaired, even though other signs and symptoms of hypothyroidism are absent. With overtreatment craniosynostosis can occur and, in addition, animal studies suggest a toxic effect on brain development. Maintenance of T_4 levels in the upper half of the normal range for age is recommended.

Initially, raised thyrotropin levels decrease with appropriate thyroid hormone replacement but may take